CASE REPORT

Intramedullary arachnoid cyst in an adult: Case report and review

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ABSTRACT

Arachnoid cysts in the spine are a rare entity with extradural occurrence being the commonest. Arachnoid cysts in intramedullary location are sparingly reported in elderly. We herein report a case of intramedullary arachnoid cyst in an adult female who presented with features of compressive myelopathy.

Key words: Arachnoid cyst, intramedullary, spinal

Introduction

Arachnoid cysts are a rare entity in the spinal cord, presenting as benign lesions usually passing off asymptomatically. Clinical manifestations appear on compression of cord or roots and consecutive neurologic symptoms. The arachnoid cysts in the spine have been classified by Nabors *et al*.[1] as: Type 1-extradural cysts without spinal nerve roots; Type 2-extradural cysts with spinal nerve root fibers; Type 3-intradural cysts. The extradural cysts are thought to arise from the herniation through the dural defects whereas the intradural cysts are thought to arise from modifications of arachnoid trabeculae. The pathogenesis of intramedullary cysts is unclear with various hypothesis of congenital, traumatic and inflammatory causes.[2-5] Intramedullary arachnoid cysts presenting with neurologic deficits have been reported in pediatric age group with only 2 case reports in adults in the literature.[6,9]

Case Report

This 54 year old female presented with dull mid-backache for 1 year duration and insidious onset, progressive weakness of both lower limbs and paraesthesias for duration of 2 months. She had difficulty in holding the footwear on which slipped out, with her knowledge suggestive of intact posterior column sensations. There was no history of bowel and bladder disturbances. She had no history of trauma in the past. On examination, she had normal upper limbs with spastic weakness (MAS-1+) of both lower limbs with power of 4/5 in all major groups and she required support to walk. There was no difficulty in getting up from supine to sitting posture. Her lower limb reflexes were brisk with bilaterally upgoing plantars. She has decreased sensations to pin prick, touch and temperature below T12 with spared sacral sensations.

Her routine hematological and biochemical investigations were within normal limits. MRI of the spine showed an intra axial lesion, hypointense on T1 [Figure 1 a and c] and hypeintense on T2 [Figure 1b and d], extending from T9-T11 region [Figure 1a and b], with no contrast enhancement [Figure 1e and f]. There was no evidence of any communication with the central canal. Preoperatively we thought it to be a syrinx or cystic teratoma.

Operation

The patient underwent T8 to TD12 laminectomy. Following durotomy, the cord was found to be swollen. The cyst was tapped followed by midline myelotomy. There was clear fluid like CSF. A thin membrane was found and was biopsied and sent for histochemical analysis. There was no communication of the cyst to the central canal which was cross checked by absence of CSF leak on Valsalva maneuver. A watertight dural closure was performed.

In the post operative period the patient developed pseudomeningocele which was managed with placement of lumbar drain. The swelling gradually subsided. She was discharged on 10th postoperative day with normalization of tone and muscle strength. Paraesthesias had significantly reduced. On regular followup there paraesthesias disappeared completely at one year. Histopathological examination revealed a single layer of meningothelial cells with fibrous

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tissue suggestive of arachnoid cyst [Figure 2]. Post operative MRI after one year showed complete disappearance of the lesion [Figure 3a and b].

Discussion

Spinal arachnoid cysts are rare, with the occurrence of intramedullary arachnoid cysts being the least as compared to the intradural extramedullary and extra dural arachnoid cysts. They are usually asymptomatic, but become symptomatic once the cyst starts compressing the cord or nerve roots. The most common presenting symptom is slowly progressive weakness in the limbs because of gradual and continuous enlargement of the cyst. MRI demonstrates the extent, size and nature of the cysts. The arachnoid cysts have similar intensities as that of CSF on T1 and T2 weighted images. T2-weighted MRI demonstrates heterogeneous signal intensity, depending on the flow effect in the cyst fluid. The imaging differential diagnosis includes-neurentric cysts, cystic teraromas, cysts associated with hemangioms, secondary to spinal tumors, post inflammatory cysts and post traumatic cysts.

The origin of intramedullary arachnoids cysts is not yet well defined. Aithala *et al.*[7] described the first arachnoid cyst in intramedullary location. Voss[8] and Rabb[9] had described arachnoid cysts in relation with dysraphic anomalies of spinal cord. Gelabert *et al.* described the occurrence of intramedullary cyst without neural tube defects.[5]

Fortuna and Mercuri hypothesised that intramedullary arachnoid cysts arise as secondary cystic development of the atypical

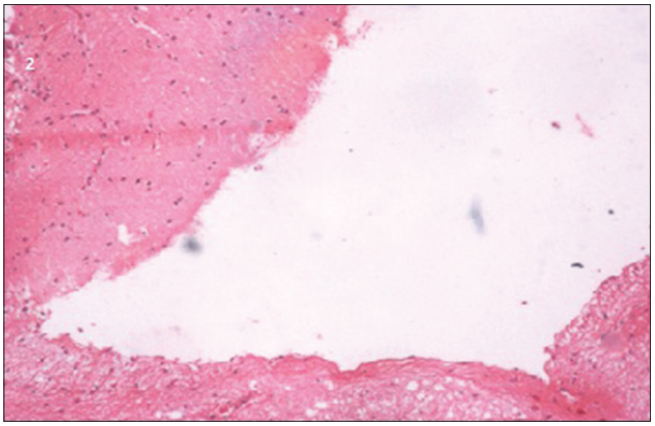

Figure 2: Histopathological examination revealed a single layer of meningothelial cells with fibrous tissue suggestive of arachnoid cyst

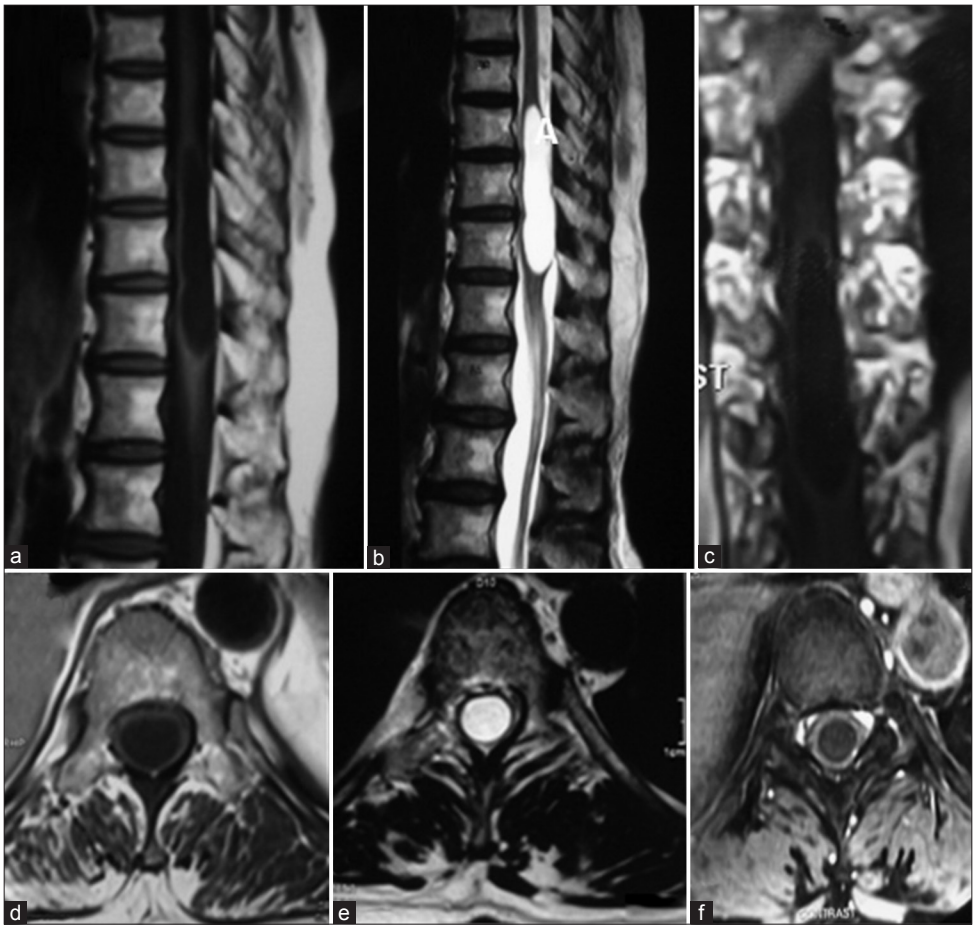

Figure 1: MRI of the spine showed an intra axial lesion, hypointense on T1 (a and c) and hyperintense on T2 (b and d), extending from T9-T11 region (a and b), with no contrast enhancement (e and f)

Table 1: Literature review of patients with intramedullary arachnoid cyst

Author and year	Age/sex	Clinical findings	Cyst location	Out come	Follow up
Aithala GR *et al.*,1999[7]	7 yrs/M	Neck rigidity, abdominal pain, paraparesis	D4	Improved	-
Gilabert-Gonzalez *et al.*, 2001[5]	12 months/F	Progressive paraparesis	L1-S1	Improved	-
Goyal *et al.*,2002[11]	63 yrs/F	Paraparesis, Incontinence	D9-L2	Improved	3 months
Sharma *et al.*,2004[3]	10 yrs/F	Quadriparesis	C4-D2	Improved	1 month
Sharma *et al.*,2005[4]	4 yrs/F	Quadriparesis	C4-C6	Improved	17 months
Guzel *et al.*,2006[2]	7 yrs/F	Quadriparesis	C2-C4	Partial improvement	24 months
Ghannane H *et al.*,2007[12]	4 yrs 8 yrs	Paraparesis Paraparesis	D3-D4 D3-D4	Complete recovery	-
Gezici *et al.*,2008[6]	35 yrs/F	Paraparesis and urinary incontinence	D5-D6	Improved	3 years
Lmejjati *et al.*,2008[13]	12 yrs/M	Paraparesis	D3-D4	Improved	4 months
Medved *et al.*,2009[14]	18 m/M	Paraparesis, constipation, neck rigidity	D5-D6	Complete recovery	1 month
Present case	54 yrs/F	Paraparesis	D8-D11	Improved	12 months

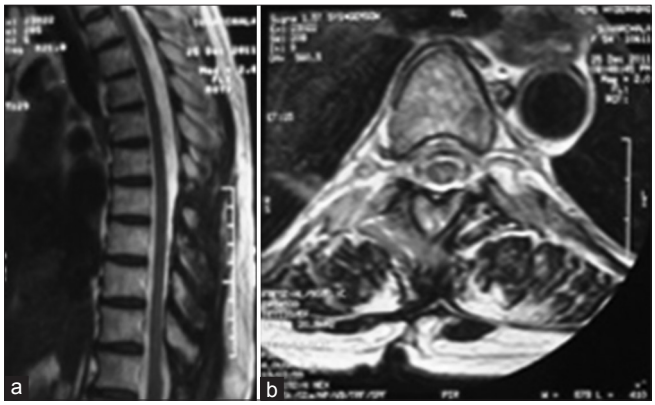

Figure 3: Post operative MRI after one year showed complete disappearance of the lesion (a and b)

intramedullary arachnoids granulations that become trapped in various locations with consequent CSF production and hence, cyst formation.[10] This view was also seconded by Goyal *et al.*[11]

Alternative hypothesis postulated describes anatomical communication between the cyst and subarachnoid space as one way valve allowing the CSF to seep into the cyst and consequent expansion of cyst.[5] In our case there was no obvious communication between the cyst and the spinal canal which was evident by no CSF leak on Valsalva maneuver.

The treatment consists of decompression of the cyst and safe removal of the cyst wall as much as feasible avoiding neurological deficits. Marsupilisation of the cyst wall is an acceptable option communicating the cyst to the subarachnoid space.

The pathogenesis of the cyst and its manifestation in adults is beyond the scope of this article.

As seen from the above published data, it is evident that even the intramedullary variant of arachnoids cysts are more common in the thoracic cord and the affected patients were mainly in the paediatric age group, more commonly in the first decade.[4,6,7,9-11,13] Whereas the two case reports published by Goyal *et al.*[11] and Gezici *et al.*[6] the affected patients were of 63 year and 35 year old respectively. Our 54 year old female patient is the third case of symptomatic intramedullary arachnoid cyst in the adult age group.

In all the reported cases surgical decompression was treatment of choice and all patients showed improvement post operatively [Table 1]. In our patient we have done a median myelotomy, decompression of the cyst and partial cyst wall excision in view of its adherence to the cord. In the post operative follow up for 12 months she improved symptomatically, with her power being improved to 5/5 in both lower limbs. Post operative MRI showed complete resolution of cyst.

Conclusion

Spinal intramedullary arachnoid cyst though rare should be considered as a differential diagnosis even in adults and surgical decompression affords good clinical results.

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